Spinraza™ (nusinersen)

Medical Guideline Disclaimer

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Definitions

Spinraza is a survival motor neuron-2 (SMN2)-directed antisense oligonucleotide indicated for the treatment of spinal muscular atrophy (SMA). The drug is administered intrathecally.

Spinal muscular atrophy (SMA) is neurological disease characterized by loss of motor neurons in the spinal cord and lower brain stem, resulting in severe and progressive muscular atrophy and weakness. 5q-SMA is an autosomal recessive genetic disorder caused by mutations in the SMN1 (survival motor neuron) gene that is found on chromosome 5. To develop SMA, an individual must inherit two faulty SMN1 genes, one from each parent.

- SMA Type 1 (infantile onset SMA or Werdnig-Hoffmann disease) — symptoms are present at birth or by the age of 6 months
- SMA Type 2 — onset of symptoms between the ages of 7 and 18 months and before the child can stand or walk independently
- SMA Type 3 — onset of symptoms after 18 months, and children can stand and walk independently, although they may require aids
- SMA Type 4 (adult-onset SMA or Kugelberg-Welander disease) — onset of symptoms in adulthood, and people are able to walk during their adult years.

Dosing and Administration

Spinraza Package Insert

Guideline
Spinraza is considered medically necessary for the treatment of Types I, II or III SMA in pediatric and adult patients when the following criteria are met.

A. Initiation therapy; all:

1. Diagnosis of SMA by, or in consultation with a neurologist with expertise of SMA
2. Spinraza is being prescribed by, or in consultation with a neurologist with expertise of SMA
3. Clinical documentation of 5q SMA homozygous gene mutation, homozygous gene deletion or compound heterozygote (i. or ii, AND iii.)
   i. Homozygous gene deletion or mutation (e.g., homozygous deletion of exon 7 at locus 5q13)
   ii. Compound heterozygous mutation (e.g., deletion of SMN1 exon 7 [allele 1] and mutation of SMN1 [allele 2])
   AND
   iii. Patient has Documentation of genetic testing confirming at least 2 copies of SMN2
4. Baseline exam of at least ONE of the following exams to establish baseline motor ability:
   i. Hammersmith Infant Neurological Exam (HINE) (infant to early childhood)
   ii. Hammersmith Functional Motor Scale Expanded (HFMSE)
   iii. Upper Limb Module (ULM) Test (Non ambulatory)
   iv. Children’s Hospital of Philadelphia Infant Test of Neuromuscular Disorders (CHOP INTEND)
5. Patient is NOT dependent on either invasive ventilation or tracheostomy, OR Non-invasive ventilation for at least 12 hours per day
6. One of the following:
   i. Patient has not previously received gene replacement therapy for the treatment of SMA; or
   ii. Both of the following:
      i. ♦ Patient has previously received gene replacement therapy; and
      ii. ♦ Patient has experienced a declination in clinical status that represented a potential failure or abatement of gene therapy efficacy
7. Spinraza is to be administered intrathecally by, or under the direction of, healthcare professionals experienced in performing lumbar punctures
8. Spinraza dosing is in accordance with the United States Food and Drug Administration approved labeling: maximum dosing of 12mg for each loading dose
9. Initial authorization will be for no more than 4 loading doses

B. Continuation therapy; all (1–5):

1. SMA diagnosis and treatment prescription by, or in consultation with a neurologist
2. Patient continues to meet ALL of the initial criteria requirements outlined above
3. No respiratory dependency on either:
   a. Invasive ventilation or tracheostomy
   b. Non-invasive ventilation for a period ≥ 6 hours per day
4. Prevention of permanent ventilation (≥ 16 hours ventilation/day continuously for> 21 days in absence of an acute reversible event or tracheostomy)
5. Clinical documentation delineates positive therapeutic response to Spinraza, from pretreatment baseline, as demonstrated by any of the measurement tools (a, b, c, or d, as appropriate) (Physician evaluation must occur ≤ 1 month prior to request)
   a. Hammersmith Infant Neurological Examination (HINE) milestones (for infants 2 months–2 years of age (i and ii):
      i. One of the following:
         1. Improvement, or maintenance of previous improvement, of at least 2 point (or maximal score) increase in ability to kick
         2. Improvement, or maintenance of previous improvement, of at least 1 point increase in any other HINE milestone (e.g., head control, rolling, sitting, crawling, etc.), excluding voluntary grasp
      ii. One of the following:
         1. Improvement or maintenance of previous improvement in more HINE motor milestones than worsening, from pretreatment baseline (net positive improvement)
         2. Member achieved and maintained any new motor milestones that is otherwise not expected (e.g., sit unassisted, stand, walk)
   b. Hammersmith Functional Motor Scale (HFMSE): (i. or ii.)
      i. Improvement, or maintenance of previous improvement, of at least a 3 point increase in score from pretreatment baseline
      ii. Member has achieved and maintained any new motor milestone from pretreatment baseline that is otherwise not expected
   c. Upper Limb Module (ULM): (i. or ii.)
      i. Improvement or maintenance of previous improvement of at least a 2 point increase in score from pretreatment baseline
      ii. Member has achieved and maintained any new motor milestone from pretreatment baseline that is otherwise not expected
   d. Children’s Hospital of Philadelphia (CHOP) infant Test of Neuromuscular Disorders (INTEND): (i. or ii.)
      i. Improvement, or maintenance of previous improvement, of at least a 4 point increase in score from pretreatment baseline
      ii. Member has achieved and maintained any new motor milestone from pretreatment baseline that is otherwise not expected

Quantity Limit:
- Initial: 4 vials for the first 58 days
- Maintenance: 1 vial every 120 days

Duration of Approval:
- Initial: 2 months
- Renewal: 12 months

Limitations/Exclusions
Spinraza is not considered medically necessary for any indication other than as listed above.
*EmblemHealth considers Spinraza not medically necessary for individuals in current treatment or previously treated with gene therapy (e.g. Zolgensma) for SMA.

Applicable Procedure Codes

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<th>Code</th>
<th>Description</th>
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<td>J2326</td>
<td>Injection, nusinersen, 0.1 mg (Eff. 01/01/2018)</td>
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<tr>
<td>J3490</td>
<td>Unclassified drugs (prior to 1/1/2018)</td>
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<tr>
<td>J3590</td>
<td>Unclassified biologics (prior to 1/1/2018)</td>
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Applicable Diagnosis Codes

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<thead>
<tr>
<th>Code</th>
<th>Description</th>
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<tbody>
<tr>
<td>G12.0</td>
<td>Infantile spinal muscular atrophy, type I [Werdnig-Hoffman]</td>
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<tr>
<td>G12.1</td>
<td>Other inherited spinal muscular atrophy</td>
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<td>G12.8</td>
<td>Other spinal muscular atrophies and related syndromes</td>
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<tr>
<td>G12.9</td>
<td>Spinal muscular atrophy, unspecified</td>
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Revision History

8/6/2020 – Updated package insert hyperlink to reflect current PI
- Updated Guideline to include “pediatric and adult patients”
- Removed pediatric neurologist requirement, changed prescriber requirement to “prescribed by, or in consultation with a neurologist who specializes in SMA”
- Updated genetic testing requirement from “no more than 2 copies of SMN2” to “at least two copies of SMN2”
- Removed age requirement
- Added requirements around previous gene therapy
- Continuation Criteria - added statement: Patient continues to meet ALL of the initial criteria requirements outlined above
- Updated duration of initial approval from 6 months to 2 months
- Updated duration of renewal approval from 6 months to 12 months

7/18/19 – Added statement: Emblem Health considers Spinraza not medically necessary for individuals in current treatment or previously treated with gene therapy (e.g. Zolgensma) for SMA

References

Med Lett Drugs Ther. 2017 Mar 27;59(1517):50-52. Nusinersen (Spinraza) for spinal muscular atrophy.


Specialty matched clinical peer review.